165 Management of pneumothoraces in cystic fibrosis (CF): a novel approach using an ambulatory suction device (Thopaz™ system, Medela Ltd)

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Objectives: Pneumothorax in CF is associated with poor prognosis, increased hospitalisation and deteriorating lung function. Persistent air leaks and delayed lung expansion lead to slow resolution. We aimed to improve care using ambulatory suction.
Methods: 3 adults with CF were treated with an ambulatory suction device (Thopaz™ system) used with intercostal drainage.
Results:

1. Surgical emphysema and persistent air leak in a 22 year old male with pneumothorax led to change from a small bore to large bore drain with suction. Lack of ambulation and chest pain limited airway clearance. Drain blockage required repeated flushes. Thopaz system improved airway clearance, reduced pain and measured air leak. Self flushing mechanism reduced drain blockage, allowing resolution in 10 days.
2. 19 year old male with end stage lung disease developed a pneumothorax. Minimal resolution and persistent air leak were seen despite a 12F drain. Patient declined large bore chest drain and/or suction due to limited ambulation. Surgical options were considered too high risk. Use of Thopaz system allowed suction use, maintained ambulation and measured air leak. Successful resolution occurred in 7 days.
3. A 22 year old female presented with ipsiliateral pneumothorax following failed pleurodesis with persistent air leak and severe dyspnoea. End stage disease and thrombocytopenia prevented surgical treatment. Pneumothorax resolved with Thopaz system aiding symptom palliation.

Conclusion: Pneumothorax in CF is complicated by limited surgical options and slow resolution. Use of an ambulatory suction device improves outcomes and reduces patient morbidity. We aim to trial this device in future patients.

166 The prevalence and aetiology of pleural effusions and empyema in patients attending a regional adult CF centre

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Objectives: To review the prevalence and aetiology of pleural effusions and empyema in a large cohort of adult patients with Cystic Fibrosis (CF).
Methods: This was a retrospective study looking at all clinical episodes between 01/01/2008 and 01/01/2014. Data for age, weight, lung function, CRP, pleural fluid analysis, transplant status, intravenous (IV) antibiotics and Read/SNOMED codes relating to pleural disease were extracted from the unit's electronic patient care records.
Results: A total 440 patients [median age 30 (17–70) years, M: 244] were reviewed. A total of 4,242 courses (67,641 days) of IV antibiotics were administered over the six year period. Three effusions were reported in 49 post-transplant patients, the aetiology being related to rejection, post transplant surgery and a spontaneously resolving lymphocytic effusion. Two cases of loculated empyema were diagnosed in non transplanted patients chronically colonised with *P. aeruginosa* (PA). Pleural Ph, LDH, Protein, Microbiology were 7.23, 1349 iu/L, 47 g/L, culture negative and 7.019, 1236 iu/L, 49 g/L, PA positive respectively. Both patients had elevated CRP (96 and 290 mg/L) and presented with pyrexia and acute pleuritic chest pain. One case responded to drain insertion and IV antibiotics, while the other required installation of intrapleural tissue plasminogen activator (t-PA) and Dnase. Patients made a full recovery.
Conclusion: Non infective pleural effusions only occurred in post transplant patients. Despite over 4000 courses of iv antibiotics, there were only two cases of parapneumonic effusions/empyema. The MIST 2 protocol using (t-PA) and DNase proved a successful alternative to surgical intervention.

167 Epidemiology and risk factors for CF arthropathy

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Objectives: Extrapulmonary complications play an important role in aging patients (pts) with CF. From a clinician's perspective CF related arthropathy (CFA) constitutes a significant manifestation of these comorbidities, especially in pts with advanced CF disease. We conducted this trial in order to assess the prevalence of CFA and to identify predictors.
Methods: 290 pts currently under treatment in our CF center were analysed in this trial. Data were collected prospectively and by standardized documentation and then matched with data from our quality management system.
Results: 43/193 adult pts suffered from relevant arthropathy symptoms of those 16 were diagnosed with rheumatoid arthritis. No Children were affected. 12/16 of the affected pts were female, their mean age was 42 years, mean BMI 21 and mean FEV1 predicted was 51%. 75% had a chronic infection with *Pseudomonas aeruginosa* 44% had CFRD. 38% were positive for rheumatoid factor. Intermittent treatment with systemic corticosteroides and NSAID was necessary in all cases. In three pts methotrexate was used. Two pts were dependent on continous therapy.
Conclusions: Our data suggest that in comparison to the general population rheumatoid arthritis' prevalence is eight fold higher in adult pts with CF leading to a significant limitation in quality of life. Pseudomonal colonisation may be a risk-factor. Confirmation of these findings in larger cohorts is required. Further research could feature ultrasound, X-ray, questionaires, screening for surrogat parameters such as rheumatoid factor and others, elucidation of immunologic mechanisms as well as the assessment and development of therapeutic strategies.

168 Cystic fibrosis: an ageing condition

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Objectives: Advances in therapy have improved survival in CF (predicted UK median now 43.5 years), but better disease awareness means that adult diagnoses are frequent, potentially skewing the survival rate. We looked at the disease characteristics of our older (>44 years) patients, and compared adult (AD) diagnoses (58%) with those in childhood (CD).
Results: 8.45% (24) of 284 patients are >44 years (50% male). All have chronic sputum infection (96% *P. aeruginosa*, 13% *Burkholderia* spp., 25% *S. aureus*, 13% MRSA, 17% NTM). The mean [SD] FEV_1 is 61% predicted [26] (CD 56% [14], AD 64% [29], P = NS) and the mean BMI 26.5 [4.8] (CD 24.8 [3.3], AD 27.8 [5.2], P = NS). 17% take oxygen therapy and 8% await lung transplant. Comorbidities include hiatus hernia, Barrett's oesophagus, splenomegaly, hypertension, angina, anxiety, constipation and colonic cancer. Common complications are shown in Table 1 and social status in Table 2.

Table 1. Common complications

	CD	AD	Total (%)		CD	AD	Total (%)
CF-related diabetes	7	6	13 (54.2)	Asthma	1	6	7 (29.2)
Liver disease	4	2	6 (25)	GORD	9	13	22 (91.7)
Cholecystectomy	4	0	4 (16.6)	Depression	2	2	4 (16.6)
Fatty/atrophic pancreas	9	1	10 (41.7)	Arthropathy	2	3	5 (20.8)
ABPA	2	5	7 (29.2)	Osteopenia/ osteoporosis	4	5	9 (37.5)

Table 2. Social status

Marital status	n (%)	Employment status	n (%)
Married or long-term relationship	20 (83%)	Full time/Housewife	9 (38%)/2 (8%)
Divorced	2 (8%)	Part time	3 (13%)
Single	2 (8%)	Retired	10 (42%)

Conclusion: The ageing CF population have more CF related complications and increased co-morbidity. Their physical characteristics are independent of age at diagnosis, reflecting the holistic approach offered by adult CF centres where close communication with other specialities is required to ensure best possible care.